Persistence of fetal circulation in the newborn

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Recently the so called syndrome of persistence of the fetal circulation has been discovered to be an important cause of cyanosis in newborns. Persistence of fetal circulation is defined as a prolongation of the transitory pulmonary vascular obstruction in the neonate, resulting in pulmonary hypertension and right-to-left shunting through the patent ductus arteriosus and the patent foramen ovale in the absence of cardiac malformations and respiratory distress syndrome.

PFC - syndrome : patients data

patients	sex	birth weight (g)	gestational age (weeks)	history
1 Z.C.	f	1.200	29	idiopathic
2 K.H.	f	2.730	37	aspiration
3 S.L.	f	4.350	40	cytomegalie
4 R.S.	m	2.500	39	aspiration
5 K.D.	m	3.800	41	aspiration
6 O.A.	f	3.060	38	EPH-gestosis
7 E.M.	m	3.240	38	aspiration

f: female, m: male

From the etiological point of view it is convenient to separate two different types. The idiopathic type of the disease is less common and usually not associated with any other systemic disorder. Persistence of the fetal circulation associated with individual cases of severe aspiration syndrome, perinatal asphyxia, bacterial or viral infections and hyperviscosity syndrome characterizes a second type of the disease.

Within a one-years-period we observed PFC-syndrome in 7 out of 345 neonates requiring intensive care (Table 1). There was the typical onset of symptome within the first 12 hours of life in all our 7 patients. Striking generalized cyanosis and apical late systolic cardiac murmurs where an uniform pattern in all cases, mimicking congenital cyanotic heart disease. On the other hand predominant pulmonary diseases had to be considered because

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of significant tachypnoe and radiological signs of parenchymal pulmonary disease in 6 patients. Primarily polycythemia, acidoses and hypoglycemia should be excluded or treated when identified.
An important clue to the diagnosis of persistent fetal pathways is the demonstration of extrapulmonary venoarterial shunting by the differential hyperoxia test (Table 2). Higher values of arterial PO_2 in the ascending aorta compared to the descending aorta evaluate right-to-left shunting through the patent ductus arteriosus. Both values, however, usually do not exceed 60 - 65 mm Hg.

PFC - syndrome : laboratory findings

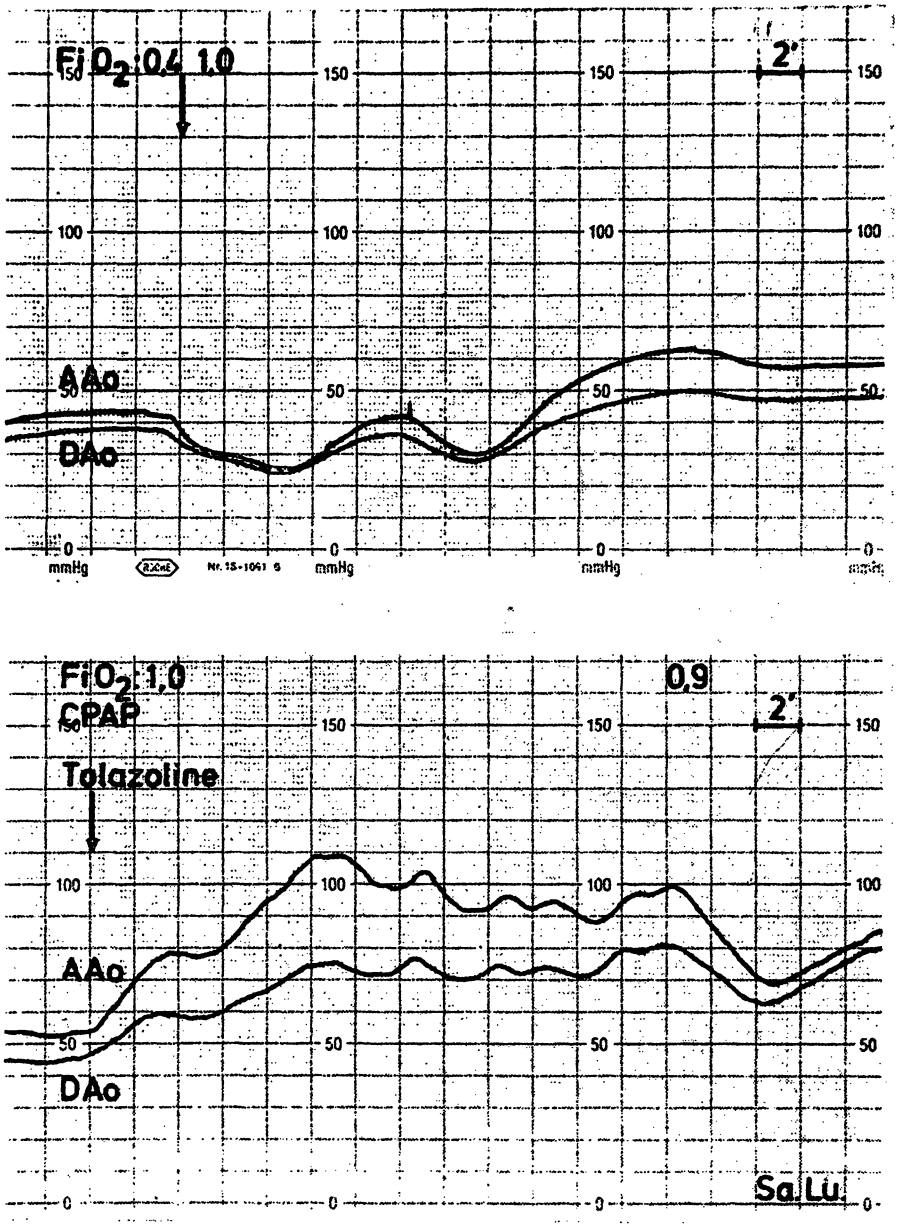
patient	Hct %	pH mm Hg	pCO_2 mm Hg	pO_2 a.a.	mm Hg d.a.
1 Z.M.	37	7,23	49	55	25
2 K.H.	37	6,85	110	58	(23)
3 S.L.	54	7,30	30	60	40
4 R.S.	45	7,16	86	35	(24)
5 K.D.	51	7,32	141	36	(24)
6 O.A.	47	7,39	44	37	(10)
7 E.M.	40	7,19	55	49	42

a.a. : ascending aorta

d.a. : descending aorta

( ) : capillary samples

In our experience transcutaneous PO_2 monitoring has proven to be as direct arterial blood sampling. Figure 1 shows an original tracing of simultaneous transcutaneous PO_2-measurements from the upper chest and lower limb. There is only a neglectible difference while breathing 40% of oxygen. Following a ten minutes breathing of 100% oxygen $P_{ct}O_2$ demonstrates only a small absolute rise in both curves but a significant increase in their difference.
At this point the differential diagnosis still encludes cardiac malformations with a ductal right-to-left shunting such as coarctation of the aorta of the preductal type or interrupted aortic arch. If these cardiac lesions are suspected and cannot be excluded by clinical means cardiac catheterization may be necessary.
Before considering an invasive procedure it may be tried to decrease pulmonary vascular resistance by a test dose of



tolazoline a potent vasodilator. Increased pulmonary perfusion immediately results in a better arterial oxygenation of the blood deriving from the left ventricle - upper curve - . The admixture of this higher arterial blood to the venous ductal blood also increases the PO_2 in the descending aorta - lower curve - however to a lesser degree.
It must be pointed out that tolazoline also decreases systemic resistance. Severe arterial hypotension from which some patients have died has been reported in the literature. Before the administration of tolazoline therefore special contraindications must be considered. Dosis regimen used, and contraindications are shown in Fig.2.

PFC - syndrome : tolazoline treatment

contraindications :

- known or suspected cardiac malformation
- arterial $pO_2 > 60$ mm Hg or $FIO_2 = 1,0$
- congestive heart failure
- systolic arterial blood pressure < 60 mm Hg

dosage :

bolus injection : 1 mg/kg within 30 sec

continuous infusion : 1 - 2 mg/kg/hour into an arm or a scalp vein

Reviewing our cases it can be summarized:

- all infants presented as critically ill, with central cyanosis and respiratory insufficiency required artificial ventilation.
- differential hypoxia test revealed significant right-to-left shunting through the patent ductus arteriosus.
- in some cases congenital heart disease can only be excluded by invasive diagnostic.
- tolazoline infusion may cause dramatic improvement in the idiopathic type and in selected cases of the secondary type.
- to avoid cerebral complications or even death long standing hypoxemia which may be responsible for cerebral convulsions in our cases it seems to be necessary to start treatment as early as the diagnosis can be established.

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